

Winter-Spring 2001

In This Issue —

- ✂ *Currarino's Triad*
- ✂ *Staged Embolization of AVM*
- ✂ *Historical Note: Dr. George Hayes*
- ✂ *Expansion of Support Personnel*



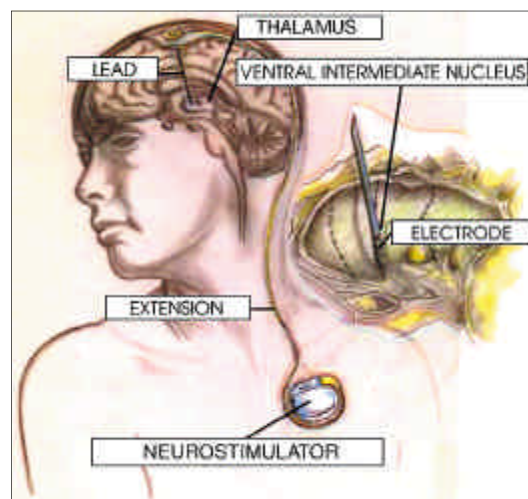
While serving as an assistant sanitary officer in New York City, Walter Reed was upset by the ignorance of the leading physician in the local community. The physician had arrived to provide information for an official report of the death of a pediatric patient. He had all the trappings of a materially successful practice including fine clothing and a splendid carriage attended by his personal footman. However, Dr. Reed later confided in his brother, Christopher, that the physician was unable to describe the child's symptoms or even to express himself in proper medical language. Dr. Reed declared the physician "a first class quack" and cried in disgust, "I shall leave the profession!" Christopher consoled him—at least for the moment.

Surgical Therapy of Parkinson's Disease

Parkinson's Disease (PD) is a progressive neurodegenerative disease of the basal ganglia affecting approximately 500,000 people in the United States. The disease is characterized by a triad of symptoms: bradykinesia (slow movement), rigidity (stiffness), and tremor. Simple tasks such as getting up from a chair, walking, and writing become increasingly difficult over months and years. In the 1960's levodopa was discovered to largely improve these symptoms, and that medication is still a cornerstone of medical therapy for PD. Unfortunately, however, over time many patients have less beneficial effects from levodopa containing drugs and they can also develop severe side effects from the medication. Drug-induced dyskinesias (abnormal movements) are a major problem for patients. Likewise, patients have fluctuations in their response to the medications. They develop "on" states when the medicine is effective and "off" states when the medication does not treat their symptoms.

Because of the limitations of medical therapy, a great deal of research has gone into developing surgical therapies. Targets for surgery include the thalamus, the globus pallidus, and the subthalamic nucleus. Initially, most surgeries were ablative: that is, a lesion was made in the appropriate target to destroy the neurons causing symptoms. Although results were often somewhat favorable, the procedures had serious limitations, especially when both sides were treated.

In the last few years most efforts have been devoted to studying the effects of high-frequency stimulation of those CNS targets. A four-contact electrode is implanted into a chosen target in the brain with the help of stereotactic localization and sometimes electrophysiological localization as well. An extension wire is then tunneled under the skin to the chest area and is hooked up to a generator that is implanted under the skin similar to a cardiac pacemaker. A specially trained neurologist then programs the device to



A neurostimulator is implanted subcutaneously and attached to an electrode implanted in one of the target centers in the basal ganglia.

—Illustration courtesy of Medtronic, Inc.

Unexpected Retorts

Repairing Bad Parts

The professor of neurosurgery watched as a mechanic removed the head of the engine of his Jaguar automobile to grind the valves.

Noticing the professor's attention, the mechanic turned to him and said, "You know, doctor, I sometimes believe this type of work is as complicated as the work you surgeons do."

"Perhaps," the neurosurgeon replied, "but the big difference is I do it when the engine is still running."

Military Medicine?

"What kind of doctor are you?" asked the inquisitive airline passenger who noticed the uniformed officer seated next to her was reading a medical journal.

"I'm a Naval surgeon," he replied with pride.

"Goodness," said the lady, "how you doctors do specialize these days!"

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Historical Note

George J. Hayes, M.D.

Second Program Director of Neurosurgery Residency

George Joseph Hayes was born on July 10, 1918, in Washington, D.C. He graduated with a B.S. degree from Catholic University of America in 1940 and received his M.D. degree from Johns Hopkins University in 1943. After completing a surgery internship at Johns Hopkins Hospital in 1944, he served as a neurosurgery resident at Lahey Clinic in Boston from 1944 through 1946. He completed a fellowship in neuropathology and neurophysiology at Duke University Hospital from 1949 till 1950 and took further training at Georgetown University Hospital in 1951. He received certification from American Board of Neurological Surgery in 1952.

Dr. Hayes began his military career as a first lieutenant in February, 1946. He served as chief of the neurosurgery service at Walter Reed General Hospital from 1947 through 1949 and again in 1950 and 1951. In 1952 he served as commanding officer of the 160th Neurosurgical Detachment in Korea before assuming command of the 46th Surgical Hospital in 1952 and 1953. He was then transferred to Brooke General Hospital as chief of the neurosurgery service from 1953 through 1955.

During and after the medical retirement of Dr. John Martin, the burgeoning neurosurgery residency at Walter Reed fell under the able caretaker administration of Drs. Harry Steelman, Martin Fefferman, and Robert Hayne. Dr. Hayes was called back to head the training program in 1955, and he continued as Chief of the Neurosurgery Service till 1966 when he was promoted to Brigadier General. Until his retirement in 1974 as a major general, Dr. Hayes served tours in the Office of the Surgeon General, the U.S. Army Medical Command in Japan, and in the offices of both the Secretary and Deputy Assistant Secretary of Defense. From 1974 till 1979 he worked as a consultant on a variety of projects dealing with civilian health matters. From 1979 through 1986 he served as medical director for Amtrak.

General Hayes has been a member of several professional organizations and held academic appointments at Georgetown, George Washington, and Baylor Universities. He published over fifty papers and received a number of honors in Korea and the United States. His wife, Catherine Conger Hayes, and he raised nine children. Avocational interests have included military medical history, archeology, photography, small arms, and drama. He lives in retirement just outside Washington, D.C.



George J. Hayes, M.D.

deliver electric stimulation to the specified area. The electric stimulus can be modified in a number of ways (intensity, frequency, etc.) to yield the best effect.

The two targets that address all the symptoms of PD are the globus pallidus (GPi) and the subthalamic nucleus (STN). The thala-

Purpose: Lead delivers electrical stimulation to a targeted site in the brain

Length: 28 and 40 cm (additional lengths available)

Diameter: 1.27 mm

Connector: In-line

Stimulating electrodes

Number: Four

Shape: Cylindrical

Material: Platinum/iridium

Contact size: 1.5 mm

Spacing: 0.5 or 1.5 mm

Materials

Insulation: Fluoropolymer

Jacket tubing: Polyurethane

Conductor wires: Platinum/iridium

Proximal connector: Nickel alloy

Stylet: Tungsten

Conductor resistance : <100 ohms

Medtronic DBS™ Lead Specifications

mus is an excellent target for the control of tremor, but most PD patients are limited more by their stiffness and slow movement than their tremor. The other benefit of stimulation is that medication can often be reduced or in some cases even stopped. The STN appears to be the target of choice right now as it treats all the symptoms very effectively and is slightly easier to target from a technical perspective. Research is continuing into the identification of other targets and for the optimal stimulation parameters for the current targets.

At Walter Reed Army Medical Center, referrals for potential candidates for electrode implantation for movement disorders are LTC K. Cannard, Chief of Movement Disorders in the Department of Neurology and LCDR L. Mulligan, Chief of Epilepsy and Functional Neurosurgery for the Neurosurgery Service.

—Submitted by Dr. Lisa Mulligan.

Staged Embolization and Microsurgical Resection of Cerebral Arteriovenous Malformations

A 49-year-old, right-handed man developed grand mal seizures in 1989 and was found to have a cerebral arteriovenous malformation. However, not until he presented for replacement of his aortic valve because of symptomatic stenosis, did he elect to undergo definitive treatment of his AVM prior to valve replacement. Physical examination demonstrated a holosystolic III/VI cardiac murmur but no neurological deficits of cognitive or sensorimotor function. Echocardiogram revealed an aortic valve diameter of 0.3 cm² associated with a pressure gradient of 45 mm Hg and an ejection fraction of 55%.

Neuroradiologic studies confirmed a high-flow arteriovenous malformation within the right rostral cingulate gyrus and superior corpus callosum measuring 3.5 cm by 2.5 cm by 3.0 cm. No recent hemorrhage was present. Cerebral angiography showed that the blood supply of the AVM was derived primarily from the right pericallosal and callosomarginal arteries (Figure 1A-B) with both superficial and deep venous drainage into the superior sagittal sinus and the vein of Galen, respectively. The vascular images found no associated arterial, intranidal, or venous aneurysms nor venous stenosis or varices.

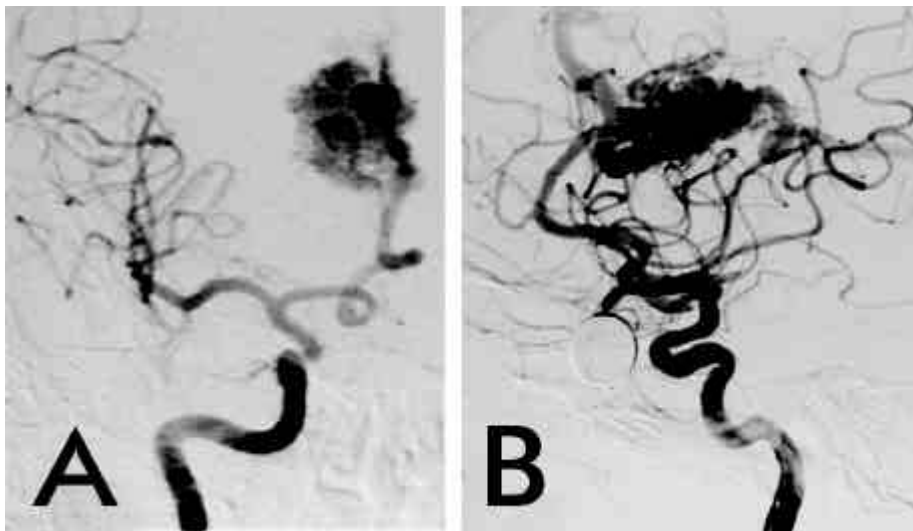


Figure 1. A high-flow arteriovenous malformation within the right cingulate gyrus and corpus callosum is fed from the anterior cerebral artery.

Owing to the patient's critical aortic stenosis and fixed cardiac output, he was advised to undergo staged embolization of the cerebral AVM followed by balloon angioplasty of the aortic valve. After angioplasty of the valve had reduced the aortic pressure gradient, he was to undergo another embolization of the AVM followed by microsurgical resection of the vascular lesion. Finally, he was advised to have the aortic valve replaced.

In order to limit the risks inherent in repeated general anesthesia in patients with

severe aortic stenosis, our patient underwent two embolizations of his AVM with medical-grade N-butyl-cyanoacrylate while awake. Thereupon, balloon angioplasty of his aortic valve was successfully completed. After the third embolization of the AVM, five arterial feeders had been obliterated, both reducing the size of the vascular nidus and markedly slowing the superficial venous drainage. Once venous slowing was evident, operative resection was scheduled for the following day.

The AVM was approached through a paramedian craniotomy that crossed the sagittal sinus. The anteriorly draining vein of the AVM had thrombosed. The pericallosal and callosomarginal arteries were dissected to gain proximal control of the blood supply. Dissection was meticulously carried out rostrocaudally along the lateral, posteroinferior, and inferior aspects of the lesion while arterial feeders were coagulated along the way. Once it was deter-

mined that there was no drainage into the posterior vein, the AVM was removed *en bloc* and confirmatory intraoperative angiography was performed.

Postoperatively, the patient developed a complete supplementary motor syndrome (see box) that nearly resolved over five days. At one month he ambulated with minor assistance and showed mild contralateral paresis of dorsiflexion of the foot as his only residual deficit.

The interdisciplinary management of this patient allowed for the safe resection of his cerebral AVM. Distal microcatheterizations and superselective angiography of the pericallosal artery in this case were accomplished by a combination of flow-guided and wire-guided techniques. Such "endovascular exploration" identified each vascular compartment of the AVM and thus permitted repeated embolizations of the lesion while sparing normal brain tissue. In suitable patients controlled hypotension under general anesthesia can aid in targeting of the embolic material within the AVM. The safety and success of embolizations is regularly assessed with electrophysiological monitoring and selective angiography.

The complexity of managing vascular malformations of the brain whether by embolization, stereotactic radiosurgery, or open resection requires neurosurgeons and interventional neuroradiologists with specialized skills and experience not widely available. Walter Reed Army Medical Center and National Naval Medical Center are fortunate in having such a team and look forward to consulting with clinicians on these difficult cases.

—Submitted by Dr. Rocco Armonda

SUPPLEMENTARY MOTOR SYNDROME

Lesions of the supplementary motor area (SMA) are commonly associated with a loss of contralateral strength, awareness, and initiation. The SMA corresponds to the area of cortex representing the mesial portion of the frontal lobe, more specifically the superior frontal gyrus. Anatomic studies have demonstrated extensive sensorimotor connections to the SMA although it is predominately motor in function with output to the primary motor cortex, cingulate gyri, contralateral SMA, spinal cord, cerebellum and basal ganglia. Its function has been defined as a protomotor cortex acting earlier than the primary motor cortex and in translation of motive to intention to action and exerting control over the primary motor cortex. It acts primarily in the preparation of internally remembered motor sequences but not in sequential activities that are sensory guided which is actually the role of the primary motor cortex. The SMA syndrome in humans has been characterized as consisting of (1) global akinesia, especially contralaterally, and speech arrest (mutism) immediately after the cerebral lesion, (2) decreased spontaneous motor activity even after recovery of a few days, and (3) the major long-term sequela being loss of rapid alternating movements of the hands.

REFERENCE — Zentner J, Hufnagel A, Perchstein U, Wolf HK, Schramm J: Functional results after resective procedures involving the supplementary motor area. *J Neurosurg* 85:542-9, 1996.





Cerebrations—*Remarks From the Program Director*



James M. Ecklund, M.D., F.A.C.S.

Welcome to the first issue of *Gray Matters* in the new millennium! As we move into 2001, our service remains vibrant, healthy, and complete. We continue to function as a single service and residency program providing full service neurosurgical specialty care at both Walter Reed Army and National Naval Medical Centers. All of our attending neurosurgeons cover call (or watch), conduct clinic, and perform operative procedures at both institutions.

For those of you who might wish to visit, our administrative offices have moved to Ward 64 in the Heaton Pavilion at Walter Reed (the “new hospital”). LTC William Monacci serves as the Army Chief of Service and helps coordinate our administrative and clinical staff at Walter Reed. At National Naval Medical Center CDR Bryan Mason has replaced CDR Robert Heim as Navy Chief of Service as Dr. Heim moved on to private practice this spring. We are delighted to have SGM (Ret.) Martin Pate join the Neurosurgical Team as our business manager this month. He recently retired as the Sergeant Major for the North Atlantic Region Medical Command and will be instrumental in ensuring continued smooth operations and enhancing our neurosurgical support throughout the region and beyond.

This spring we will launch a new initiative designed to enhance communication among DOD neurosurgeons and provide subspecialty neurosurgical consultation. We are establishing a Neurosurgical Telemedicine Case Conference available to all interested neurosurgeons in DOD. Neurosurgeons will be able to submit cases in advance over the internet, and the optimum management will then be discussed in real time during the conference. We hope eventually to expand our telemedicine operations to provide internet-based consultation to all physicians including those deployed. We are also fortunate to regularly consult with our civilian colleagues at quarterly city-wide resident case conferences where all the residents and faculty from Georgetown and George Washington join with our Program to discuss challenging cases.

This fall we were fortunate to have Dr. Ludwig Kempe return to Walter Reed to speak at the first annual Hugo V. Rizzoli Lectureship. Dr. Kempe spoke about the early days of Neurosurgery at Walter Reed General Hospital. Dr. Rizzoli was in attendance along with many former residents and faculty from both Walter Reed and Bethesda as well as George Washington University. After Dr. Rizzoli’s military service at Halloran General Hospital over 50 years ago, he remained an instrumental and dedicated consultant to military neurosurgery in the Washington, D.C., area. We were delighted to partner with the Uniformed Services University of the Health Sciences to honor Dr. Rizzoli with this annual lectureship.

Since our last newsletter we have welcomed three new attendings to our faculty. LTC Leon Moores completed a fellowship at the Children’s National Medical Center here in Washington and now directs the Pediatric Neurosurgery Section. LCDR Lisa Mulligan completed a fellowship at Yale and directs the Epilepsy and Functional Section. LCDR Marilyn L. Gates completed a fellowship at University of Wisconsin and co-directs the Spine Section.

The resident team is thriving under the leadership of the Chief Resident, MAJ Richard Gullick. The residents are involved in clinic and operative procedures at both Walter Reed and National Naval Medical Center, and are assisted in their duties by an exceptional team of physician assistants. The Residency Review Committee for Neurological Surgery recently awarded full accreditation to our newly combined Army-Navy training program. In July we look forward to having CPT Dennis Geyer join us as our next first-year resident (PGY-2), and LT Chris Neal join us as our neurosurgery intern (PGY-1). CPT Abel Jarrel has also recently joined our Neurotrauma



Dr. James Ecklund

CURRARINO'S TRIAD: Report of a Case

Currarino's triad is a group of malformations first reported by Guido Currarino and coworkers in 1981. The components of this triad are anorectal malformation, sacral defect, and presacral mass. Currarino's triad should be considered in the differential diagnosis of any pediatric patient with an anorectal malformation and any form of sacral agenesis.

Perhaps the most compelling explanation for Currarino's triad was proposed by Dias and others in 1998. Dias's group described a patient with a caudal split-cord malformation and Currarino's triad. They proposed that a failure of separation of the caudal cell mass (or "tail bud") from the hindgut during late gastrulation may be responsible for the syndrome. This theory is consistent with the anatomic presence of anterior defects of the sacral spine, since the caudal cell mass separates anteriorly from the distal hindgut. The endodermal component which forms the hindgut is distinct from the tail bud. It is the multipotent cells of the tail bud that form the caudal neural tube.

In the case below, the operative findings that include incomplete formation of the caudal somites associated with a teratoma in close approximation to the hindgut support the theory set forth by Dias. One can further speculate that the failure of separation and the resultant close association of the caudal cell mass with the hindgut induced teratoma formation.

We recently treated a 3-year, 5-month-old, Hispanic female with Currarino's triad. Shortly after birth, our patient was found to have a low, imperforate anus with an anteriorly displaced recto-perineal fistula. Corrective surgery was performed. Chest radiogram, renal and abdominal ultrasonograms, and lumbosacral MRI were reported as normal. She later developed constipation marked by hard stools passed only every three or four days. After multiple visits to the hospital for fecal impaction over the next three years, anorectal manometry was performed. The study demonstrated "some sphincter control and a large-volume rectum" but was considered within normal limits.

On physical examination the patient was a well developed a girl who was at the 50th percentile for both height and weight. Neurological findings included normal

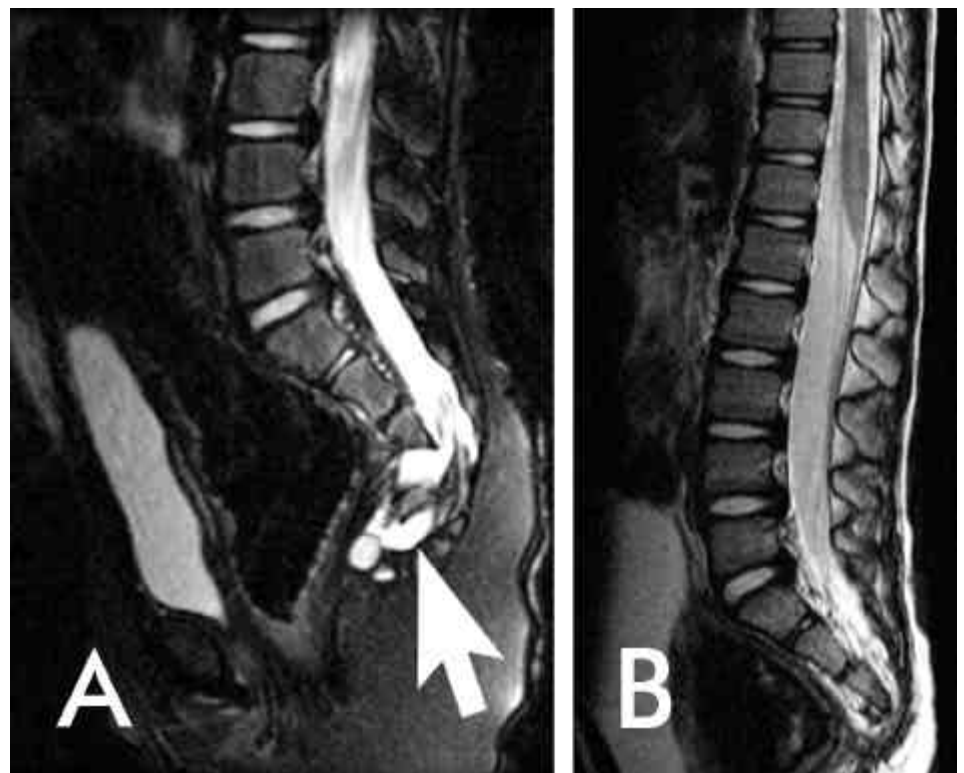


Figure 2. Currarino's triad revealed in T2-weighted magnetic resonance images before and after surgery. A. The lumbosacral spine and pelvis demonstrate an anterior sacral meningocele and small, round, hyperintense mass inferiorly (arrow). The rectum and bladder appear distended. B. The meningocele and associated soft-tissue mass have been totally resected.

lower extremity motor and deep-tendon reflexes. Sphincter tone was normal, and an anal wink reflex was elicited.

MRI of the lumbosacral spine and pelvis showed ectasia of the thecal sac and spinal canal with extension of the thecal sac through an anterior sacral defect within the pelvis, consistent with an anterior meningocele at level of S2 and S3. A 1-cm, soft tissue mass with high T1 signal and low T2 signal was also identified at the same level. The conus medullaris terminated normally adjacent to the L1 vertebra. (See Figure 2A-B)

Resection of the sacral lesions was recommended for two reasons. First, chronic constipation in this patient carried a small, but recognized risk of infectious complications such as meningitis. Second, a tissue diagnosis was needed for management of the soft-tissue mass associated with the meningocele.

The patient was taken to the operating room where a midline posterior incision was created from the L4 spinous process to the coccyx. After removal of the posterior elements of the spine with wide laminectomies, a large, anterior sacral defect was observed. Attached to the caudal end of the thecal sac was a solid mass measuring 3 cm by 3 cm by 4 cm. A midline durotomy revealed absence of spinal nerve roots below the level of S1. The thecal sac was then ligated distal to the caudalmost normal root sleeve. The soft-tissue mass was dissected free from the posterior rectum. A coccygectomy was performed, and gross total resection of the mass was accomplished. The mass appeared to be made up of multiple tissue types including fat, hair, and connective tissue. Final pathological reading of the mass was teratoma. The patient recovered well with excellent wound healing and no neurological

deficit. MRI performed three months post-operatively demonstrated no evidence of recurrent or residual mass. (See Figure 2B).

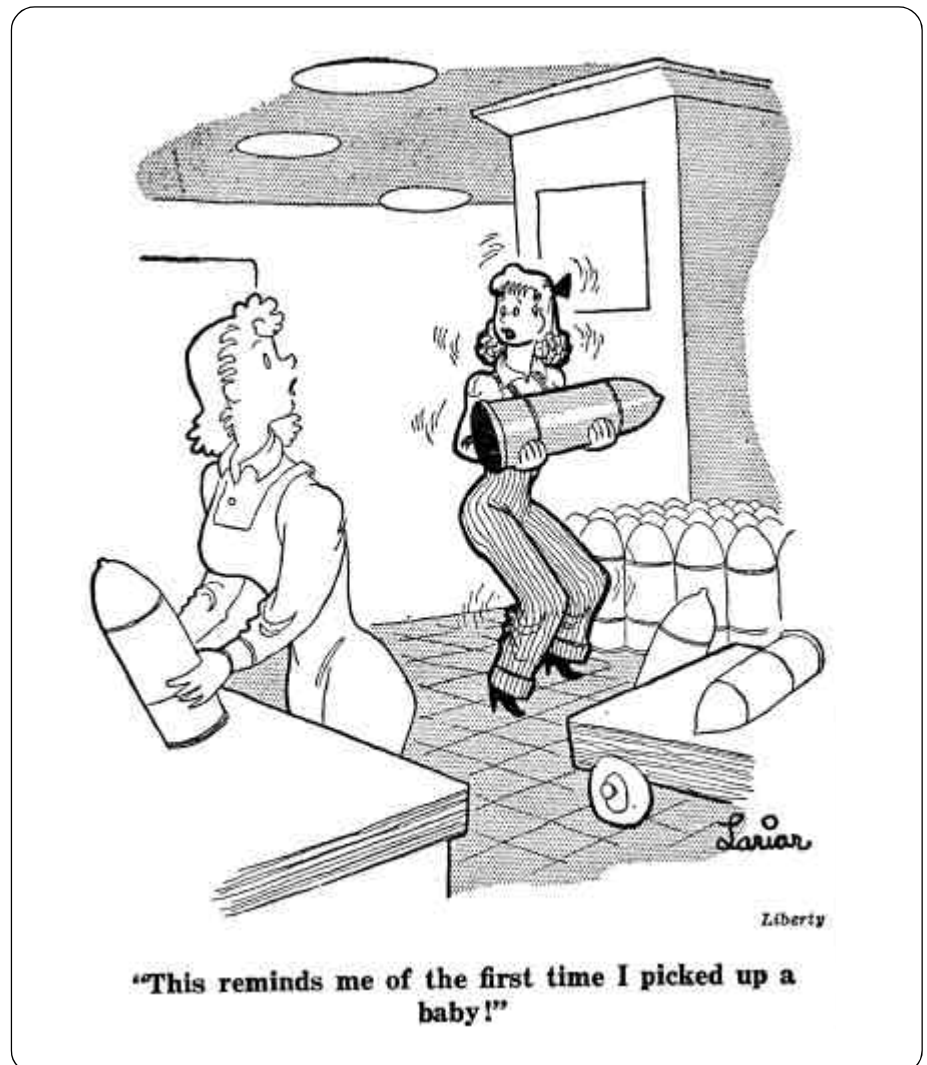
A multidisciplinary team of pediatric subspecialists from gastroenterology, radiology, general surgery, and neurosurgery permitted full preoperative diagnosis and successful management of this complex young patient with Currarino's triad. At Walter Reed Army Medical Center and National Naval Medical Center, we have a full complement of pediatric subspecialists in many other disciplines as well, and we are always available to consult with you at any time on your pediatric patients.

—Submitted by Dr. Leon Moores.

Laboratory where a number of very promising projects are underway under the leadership of LTC Geoff Ling at the medical school in Bethesda.

In September, 2000, at the annual meeting of the Congress of Neurological Surgeons, Dr. and Mrs. Lloyd Youngblood generously hosted a delightful dinner party for the faculty and alumni of our program. For those of you who attend the AANS meeting in Toronto this spring, I hope you can attend our reception on Monday evening to meet some of our current resident and attending staff, reminisce about old times, and share our enthusiasm about future opportunities.

Have an enjoyable and safe spring and summer!



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(202) 782-8756

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(202) 782-9804

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Website
[http://www.wramc.amedd.army.mil/
departments/surgery/neuro](http://www.wramc.amedd.army.mil/departments/surgery/neuro)

EXPANSION OF PROFESSIONAL SUPPORT PERSONNEL IN NEUROSURGERY

Things at Walter Reed have really changed, especially when it comes to support personnel.

In the past two years the Neurosurgery Service has evolved from a civilian staff consisting of a couple of secretaries, a receptionist, and a part-time film clerk and “man Friday” to an unprecedented complement of professionals. From a solitary neurosurgery case manager, the professional support staff has grown to three R.N. case managers, a certified nurse practitioner, three physician assistants, not to mention an additional clinical nurse specialist at Bethesda.

The availability of such staff has improved the flow and coverage of patients around the clock. Patients can be promptly screened, examined, scheduled, and managed as never before. The attending staff remain busy in the outpatient clinic and operating room. The residents have been freed of a lot of “scut” and routine paper work.

Needless to say, patient satisfaction should soon reach an all time high as the average length of hospital stays reaches an all time low!

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